Background/Purpose: Ulnar polydactylies present with a wide variation in the size and appearance of the affected finger, ranging from small cutaneous appendages to fully formed additional fingers. Since ulnar polydactyly is rarely associated with functional disabilities, psychological and cosmetic reasons are the main rationale for surgical treatment. The aim of this study was to present the appearance, treatment regimen, and outcome of a series of children treated with ulnar polydactylies.

Methods: Data of all pediatric patients treated between 2000 and 2010 were included. For assessing long-term outcome, patients were contacted via mail and asked to complete a questionnaire concerning location and appearance of the scar as well as functional and cosmetic satisfaction (VAS 0–100).

Results: In that period, 32 patients (20 male, 12 female) with 53 ulnar polydactylies were treated. In 20 patients the polydactyly occurred bilaterally (63%), in 10 patients on the left side (31%), and 2 patients presented with right-sided ulnar polydactyly (6%). The vast majority of ulnar polydactylies were floating cutaneous appendages. The mean age of the patients at operation was 8.6 months (range 0 to 10 years). 20 patients (63%) responded to the postal questionnaire. Follow-up time was 4.9 years (range 2.1 to 10 years). The majority of patients reported a flat scar (n = 18, 90%). Mean overall satisfaction with the postoperative result using a VAS 0–100 score was 89.

Conclusion: We were able to report a good postoperative outcome of a series of patients suffering from ulnar polydactylies.
were collected retrospectively and analyzed with regard to gender,
type of ulnar polydactyly, association with other hand/foot deficien-
cies, age at surgery, type of treatment and complications during
treatment. The patients’ radiographs were reviewed and the type of
ulnar polydactyly was classified according to Stelling and Turek
(Fig. 1). Additionally, bilateral radiographs of the feet were performed
in cases with clinical signs of polydactyly of the feet.

1.1. Treatment strategies

In cases of Type 1 ulnar polydactyly according to Stelling and
Turek (floating appendages) oval excision was performed under
general anesthesia. Care was taken to avoid an unesthetic amputa-
tion stump and to carry out a deep resection of the nerve in order to
prevent the development of a painful neuroma. Stelling and Turek
Type II and III ulnar polydactyly were treated with more complex
ablation of the digit including corrective osteotomy, ligament and
meticulous soft tissue reconstruction and articular contouring.
Attention was paid to preserve the tendinous and neurovascular
structures in order to facilitate optimal function of the remaining digit.

1.2. Follow up

In order to assess the long-term outcome, patients were
contacted via mail and asked to complete a postal questionnaire.
The questionnaire included questions concerning location and
appearance of the scar as well as functional and cosmetic satisfaction
(VAS 0–100). Additionally, a family history of hand and/or foot
deficiencies was surveyed.

Data were entered in a Microsoft Excel spreadsheet for
descriptive analysis.

2. Results

Between 2000 and 2010, 32 patients (20 male, 12 female) with 53
ulnar polydactyly were treated. Of these, the malformation occurred
in three pairs of siblings. In 20 patients the polydactyly occurred
bilaterally (63%), in 10 patients on the left side (31%) and 2 patients
presented with right-sided ulnar polydactyly (6%).

The vast majority of ulnar polydactyly were floating cutaneous
appendages. The exact distribution of the 53 polydactyly according
to Stelling and Turek is shown in Table 1. While 22 patients (69%)
presented with ulnar polydactyly without other anomalies, addi-
tional polydactylies of the hands or feet were recorded in 10 patients
(31%). The distribution of these additional polydactylies is presented
in Table 2.

Six patients (20%) presented with a syndrome (two with Ellis–van
Creveld Syndrome, two with Jeune Syndrome and two with an
unclassified dysmorphic syndrome) and one girl suffered from a
duodenal atresia. Of the 32 patients in our series, 13 children (40%)
had a positive family history for other types of polydactyly.

The mean age of the patients at operation was 8.6 months (range
0 to 10 years). Minor complications occurred in a total of 4 patients
(12%), of which three patients developed a hypertrophic scar and in
one patient a superficial wound infection occurred. Another patient
developed a painful neuroma necessitating a second surgical
intervention three years following the initial resection of a type I
ulnar polydactyly.

The questionnaire was sent to all 32 patients. 20 patients (63%)
responded, 3 patients (9%) did not respond due to unknown
address and 9 patients (28%) did not respond. The follow-up time
for the patients was 4.9 years (range 2.1 to 10 years) after
operation. The 20 patients who responded in total had 30 ulnar
polydactyly (n = 27 Stelling and Turek Type I, n = 3 Type II).

The majority of patients reported a flat scar (n = 18, 90%), while 2
patients stated to suffer from a residual nubbin (10%). None of
patients reported functional differences between the two hands. One
patient (5%) reported occasional pain of the scar. Mean overall

Table 1

Classification of 53 ulnar polydactyly seen in 32 patients according to Stelling and
Turek.

<table>
<thead>
<tr>
<th>Type</th>
<th>Appearance</th>
<th>Number of polydactylies</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>floating cutaneous appendage</td>
<td>41</td>
<td>77</td>
</tr>
<tr>
<td>2</td>
<td>partial duplication</td>
<td>8</td>
<td>15</td>
</tr>
<tr>
<td>3</td>
<td>complete duplication</td>
<td>4</td>
<td>8</td>
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Table 2

Characterization concerning the occurrence of other polydactyly of 32 patients
presenting with 53 ulnar polydactyly.

<table>
<thead>
<tr>
<th>Patients</th>
<th>left</th>
<th></th>
<th>right</th>
<th></th>
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</thead>
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<tr>
<td></td>
<td>ulnar</td>
<td>radial</td>
<td>foot</td>
<td>ulnar</td>
</tr>
<tr>
<td>14</td>
<td>X</td>
<td>X</td>
<td>X</td>
<td>X</td>
</tr>
<tr>
<td>7</td>
<td>X</td>
<td>X</td>
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<td>4</td>
<td>X</td>
<td>X</td>
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<td>X</td>
<td>X</td>
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<tr>
<td>1</td>
<td>X</td>
<td>X</td>
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</table>

Fig. 1. Example of a 6 month old girl with Stelling and Turek Type I ulnar polydactyly on the left side and Stelling and Turek Type II ulnar polydactyly on the right side. The patient was

treated with surgical excision of both additional fingers. At follow-up of 7.4 years the patient reported a VAS of 95, a flat scar, no pain and normal function.
A recent publication by Pritsch and coworkers describes occurrence in 40% and a bilateral occurrence in more than 60% of other reports about the Caucasian form, we found a familial satisfaction with the postoperative result using a VAS 0–100 score was 89 (ranging from 10 to 100). The detailed findings of the long-term outcome are presented in Table 3.

3. Discussion

The present study aims to assess the postoperative outcome of a series of children treated with ulnar polydactyly. Generally, ulnar polydactyly represents a common malformation of the hand. Two clinical forms of ulnar polydactyly are described in the literature, the African and the Caucasian form [11]. The African form is reported with an incidence between 1:1500 and 1:3000, most cases are sporadic, the majority of the cases are unilateral (80%) and in 10% the ulnar polydactyly is part of a syndrome [11]. Due to demographic reasons, the series of patients reported in the present study more closely resembles the Caucasian form. While we cannot provide information about the incidence, the occurrence of other malformations of the hand or foot in a third of our patients and the association with a syndrome in a fifth of the children in this series confirm these previous findings. However, compared to other reports about the Caucasian form, we found a familial occurrence in 40% and a bilateral occurrence in more than 60% of our patients. A recent publication by Pritsch and coworkers describes a positive family history of polydactyly of the foot or hand in 47% of 49 mostly Caucasian patients [8].

More than 40 syndromes are described to be associated with ulnar polydactyly including Trisomy 13, McKusic–Kauffman and Pallister–Hall syndrome [2]. The syndromal cases in the present series include Ellis–van Creveld, Jeune syndrome and two patients with an unclassified dysmorphic syndrome. The high number of syndromal cases underlines the necessity of an accurately performed examination when dealing with children seen with ulnar polydactyly.

It is widely accepted that the more complex forms of ulnar polydactyly require elaborate reconstructive procedures performed in specialized centers. Nevertheless, the majority of ulnar polydactyly are Stelling type I resembling floating cutaneous appendages [2]. Dodd and colleagues have demonstrated that there are uncertainty and inequality in treatment decision of these forms [9]. Suture ligation at the base of the duplicated digit done in the nursery represents the most commonly performed procedure [12]. The aim of this approach is to produce necrosis and subsequent autoamputation of the additional digit. Advantages of suture ligation include its cost-effectiveness and simplicity. In recent years, however, the practice of suture ligation has been questioned by various authors due to the possible development of painful neuroma or incomplete amputation leaving behind an unaesthetic scar [13]. In 2011, Patillo and Ryan reported two complications of suture ligation ablation for ulnar polydactyly. One case consisted of bilateral retained gangrene and a second case consisted of a residual sensitive skin tag [10]. When performing suture ligation the nerve of the rudimentary digit is divided at the level of the skin and it cannot retract into the soft tissue. Therefore, the development of painful neuroma is favoured. Various authors have reported patients with sensitive amputation neuromas necessitating surgical revision [14,15]. Based on these results we prefer to perform excision of Stelling and Turek type 1 ulnar polydactyly under general anesthesia with meticulous resection of the nerve.

Watson and Henrikus re-examined 15 of 21 children treated with ligation of an ulnar polydactyly at an average age of 20 months. No finger had a perfectly normal cosmetic appearance and the authors noticed a residual bump in 43% of the patients [16]. In contrary, only 2 of the 20 patients (10%) included in our study developed a bump at the excision site at a mean follow-up time of 4.9 years (see Table 3). The subjective satisfaction with the postoperative results seemed to correlate with the presence of a postoperative bump. While the two patients with the elevated scars reported a subjective satisfaction of VAS 10 and 75, the remaining patients reported a satisfaction of mean 94. These results demonstrate the superiority of the formal surgical excision of floating cutaneous ulnar digits related to postoperative patient satisfaction.

In conclusion, we were able to report the good postoperative outcome of a series of patients suffering from ulnar polydactyly. Moreover, we suggest the formal excision with meticulous resection of the additional nerve in Stelling and Turek type 1 ulnar polydactyly.

References


Table 3

Detailed findings of the long-term outcome of 20 patients treated for ulnar polydactyly.

<table>
<thead>
<tr>
<th>Occurrence</th>
<th>Stelling and Turek right</th>
<th>time of f/u [a]</th>
<th>VAS scar</th>
<th>function pain</th>
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<td>3.13</td>
<td>100 flat</td>
<td>normal no</td>
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<tr>
<td>left 1</td>
<td>4.93</td>
<td>90 flat</td>
<td>normal no</td>
<td></td>
</tr>
<tr>
<td>left 1</td>
<td>5.94</td>
<td>90 flat</td>
<td>normal no</td>
<td></td>
</tr>
<tr>
<td>bilateral 1 1</td>
<td>2.23</td>
<td>100 flat</td>
<td>normal no</td>
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<tr>
<td>bilateral 1 1</td>
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</tr>
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<td></td>
</tr>
<tr>
<td>right 1</td>
<td>2.16</td>
<td>100 flat</td>
<td>normal no</td>
<td></td>
</tr>
<tr>
<td>left 1</td>
<td>2.55</td>
<td>10 nubbin</td>
<td>normal no</td>
<td></td>
</tr>
<tr>
<td>bilateral 1 1</td>
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<td>normal no</td>
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</table>

f/u…follow-up, VAS…visual analogue scale (rated by the patients and the parents).